evaluation, pts in Grp 1 were more likely to have generalized aortic dilatation (90 vs 30%, p = 0.002), dilatation of the aortic annulus (90 vs 24%, p = 0.002), at least moderate MR (p = 0.03), at least moderate aortic regurgitation (p = 0.05), pectus carinatum (p < 0.001), and severe scoliosis (p = 0.04).

Conclusions: Predictors of adverse outcome in childhood MS include the neonatal phenotype, a family history of dissection, and generalized dilatation of the aorta at initial echo. Children with cardiovascular aortic morbidity and mortality have a high incidence of MR, AR, and severe skeletal anomalies.

2:15

814-2 Does Prenatal Diagnosis of Hypoplastic Left Heart Syndrome Lead to Improved Surgical Outcome?

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Prenatal diagnosis of Hypoplastic Left Heart Syndrome (HLHS) provides a unique opportunity to plan perinatal management. The majority of infants with HLHS are diagnosed shortly after birth, requiring resuscitation and transport to a tertiary care center. Studies from large centers report a survival of 70% after first, and 90% after second stage surgical palliation. The influence of prenatal diagnosis and elective delivery at a tertiary care center on the surgical outcome of these infants is not known. To address this question, we reviewed all patients diagnosed with HLHS prenatally and postnatally at our institution from 7/92 to 5/97. There were 61 patients diagnosed with HLHS, 25 prenatally and 36 postnatally. Of the 25 infants diagnosed prenatally, 10 (40%) underwent palliative surgery, 6 (24%) declined surgery and 9 (36%) pregnancies were terminated. Of the 36 patients diagnosed postnatally, 23 (64%) underwent palliative surgery and 13 (36%) did not. Nine out of 10 (90%) of the prenatally diagnosed infants survived first stage palliation and all (9/9; 100%) survived the second stage. In contrast, 12 of the 23 (52%) postnatally diagnosed infants survived the first stage palliation and 11 of those 12 (92%) survived the second stage. These findings suggest that prenatal diagnosis and elective delivery in a tertiary care center may contribute to improved surgical outcome through second stage palliation in patients with HLHS (90% survival vs. 48%; p < 0.05). Almost all of this improvement was at the first stage of palliation and may reflect the benefits of avoiding the hypoperfusion and acidosis commonly associated with postnatal presentation of HLHS.

2:30

814-3 Impact of Fetal (F) Echocardiography on the Course of Major Congenital Heart Disease (MCHD): A Population Based Study

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Background: Obstetric ultrasound is widely performed (94%) in NSW. Frequency of cardiac abnormality found and influence of this on outcome are examined in a comprehensive population based study.

Method: In a study of the NSW population (6 Mill; 85000 birth/y) from 1/94–12/96 we evaluated MCHD for 1) prenatal detection rate and pattern, 2) impact of F diagnosis on outcome, and 3) compared those with F diagnosis with MCHD detected after birth (≤6 mo).

Results: 99 fetuses and 563 infants with MCHD were identified (birth prevalence:0.26%; F detection rate: 15%). 4 chamber view anomalies dominated in the F group (64/99; 65%) and conotruncal lesions the postnatal (PN) group (328/563; 58%). F detection ratio (F/F + PN) was best for 3° AV block (5/6 = 83%), univentricular lesions (37/92 = 40%), 1-TGA (2/5 = 40%), and severe AV valve anomalies (19/93 = 20%), and worst for tetralogy (9/86 = 9%), large VSDs (4/75 = 5%), dTGA (0/77 = 0%), TAPVR (0/23 = 0%) and truncus (0/13 = 0%). Of the F group 34 were terminated, 11 died in utero. An intervention <2 w of age was performed in 12/45 (27%) F survivors and 213/563 (38%) of PN group. Planned intervention in the F group was achieved <2 d in all. 6 mo survival rate was 83% and 86% for the F and PN groups, with comparable outcomes for individual defects.

Conclusions: Our results demonstrate a low sensitivity of obstetric ultrasound for MCHD. F diagnosis had important implications for pregnancy, can impact favourably on perinatal management, but did not change 6 month survival of the lesions examined.

2:45

814-4 New Insight into the Single Ventricular Function After the Fontan Procedure: A Three-dimensional Echocardiographic Study

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Background: Both pulmonary and systemic circulations after Fontan operation are based on single ventricle performances. Three-dimensional echocardiography (3DE) provides volumetric data of the ventricular cavity. Our purpose was to apply 3DE after Fontan procedure to quantify ventricular volume and function.

Methods: Ten patients (median age 8 years) were studied 5 years after a Fontan operation. End-diastolic volume (EDV), end-systolic volume (ESV), stroke volume (SV) and ejection fraction (EF) of the single ventricle were calculated by 3DE and compared to the left ventricular parameters in 10 normal children matched for age, sex and size. Parameters were compared using unpaired t-test.

Results: The data are the following:

	Fontan	Controls	p
EDV (ml)	48 ± 22	64 ± 20	NS
ESV (ml)	25 ± 13	18 ± 9	NS
SV (ml)	23 ± 10	36 ± 12	0.04
EF (%)	48 ± 7	66 ± 7	0.005

Volumes of the ventricular cavity were not different in the two groups. SV and EF were lower in the Fontan patients.

Conclusions: 3DE provides a quantitative insight into the pathophysiologic function of the single ventricle after the Fontan operation.

3:00

814-5 Three-dimensional Echocardiographic Comparison of Right Ventricular Mass and Volumes Following Trans vs. Non-transannular Patch Repair in Post-operative Patients With Tetralogy of Fallot

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The hemodynamic effects of Tetralogy of Fallot (TOF) surgery have been difficult to assess due to the inability to accurately measure RV size and function. Three-dimensional echocardiography (3DE) can provide reliable volume and mass data. The purpose of our study was to 1) determine the feasibility of 3DE measurements of RV size and function in post-op TOF and 2) compare pts with non (NTP) vs. transannular patch (TP) placement at initial surgery. We performed 3DE RV volume measurements in 21 pts with TOF, med. age 11 yrs (2–42), with (n = 16) or without (n = 5) a TP and compared them with 20 controls, mean age 11 yrs (2–23). Indexed right ventricular end-diastolic (RVEDV), end-systolic volumes (RVESV), and myocardial mass (RVM) were significantly higher in the pt group. RV ejection fraction (EF) was significantly lower (Table).

TOF	MEAN	(± SD)	CONTROLS		P
RVEDV/BSA	83	(43)	47	(9)	<0.01
RVESV/BSA	47	(28)	21	(8)	<0.01
RVM/BSA	83	(47)	48	(11)	<0.01
EF (%)	45	(10)	57	(10)	<0.01

Pts with TP tended to have larger RVEDV (96 vs 50 ml/M2) and a lower EF (42 vs 55%) than NTP pts.

Conclusions: 3-DE RV mass and volume measurements can be obtained in post-op TOF pts. Post-op pts. have significantly increased RVEDV, RVESV, and RVM than normals. Transannular patch repair pts tend to have increased RVEDVs and lower ejection fractions than non-transannular patch repair pts. This preliminary experience enhances our understanding of RV size and function following TOF surgery, and encourages our efforts to attempt NTP repair when anatomically feasible.

3:15

814-6 Neonatal Coarctation of the Aorta: Is the Left Heart Ever too Small?

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Although the influence of left heart (L hrt) structure sizes on a 2 ventricle outcome for critical aortic stenosis (AS) has been described, less is known about the spectrum of L hrt hypoplasia seen with aortic coarctation (Coa). The purposes of this study were to determine 1) the spectrum and influence of L hrt hypoplasia in Coa and 2) whether the left ventricle (LV) adequacy score previously described by Rhodes for pts with critical AS is applicable to Coa. We reviewed the charts and echoes of 63 consecutive neonates (without ASD, significant AS, or complex anatomy) who underwent Coa repair between 1/93 and 4/97. Fifty had a PDA and 34 had a VSD. From the initial echo, we measured the aortic root, aortic and mitral annulus (ann), LV size and function, left to right ventricular length (LV/RV) ratio, transverse (Tx)